Anosognosia for chronic dysphasia

Clinical Postcard

ANOSOGNOSIA FOR CHRONIC DYSPHAGIA

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Aldo (not real name), a man with 17 years of formal education, was 60-years old at the time of our assessment. He was affected by a probable Progressive Supranuclear Palsy which started four years before our examination. He was treated only with antidepressants. His neuropsychological profile was characterised by executive impairments (see Table 1), which reflected in some everyday behaviour (apathy, withdrawal, social inappropriateness and fatuity). Aldo also presented with balance difficulties, clear dysarthria and severe dysphagia, characterised by altered oral and pharyngeal phases, resulting in food sticking in his throat, choking, hawking and coughing.

--- Table 1 about here---

Aldo's dysphagia was accompanied by tongue protrusion and profuse sialorrhea, causing considerable embarrassment to his wife, but apparently not to him. Curiously, he apologised when his drooling was dripping on other people or on the testing desk solely when he was perceiving negative feedback from other people. Even drinking caused substantial anterior leakage and drooling, with nasal penetration, predeglutitive aspiration and oral residues. However, on the Movement Disorder Society - Unified Parkinson's Disease Rating Scale (MDS-UPDRS – Goetz et al. 2008), section 2.2 questioning about drooling, he self-rated himself zero, i.e. no drooling. He also rated himself normal (i.e., score zero) on sections 2.3 and 2.4 of the same scale, inquiring respectively about chewing/swallowing and eating tasks, indicating a complete denial of his clear dysphagia.

On the Functional Oral Intake Scale (Crary, Carnaby Mann & Groher, 2005) measuring severity of dysphagia, he was scored 4/7, indicating that he could be fed an oral diet of a single consistency. He was referred to a speech and language therapist who instructed him (and his wife) to adopt appropriate dietary and swallowing strategies and postural adjustments, to modify food texture and to increase the viscosity of liquids.

Aldo was perfectly aware of his motor and balance difficulties, as well as of his dysarthria, for which he required insistently to be treated and exercised thoroughly following the speech therapist's advice with his wife's assistance. On a self-rating questionnaire assessing dysarthria (Schindler & Gullí, 2002) he scored 68/140, whereas both his wife and his therapist scored his problems 46/140, indicating that he slightly overestimated his articulation and language problems.

On the contrary, he strongly denied his dysphagia often arguing with his wife who worried about his frequent chocking, coughing, vomiting and breathing gasps. On the Swallowing Disturbance Questionnaire (Cohen & Manor, 2011) he scored 5/45, whereas his therapist scored his performance 28/45 and his wife 29/45, indicating a clear underestimation of his swallowing problems. For example, to the question "Do you have any difficulty when eating solid food?" he responded, "No problems at all, I am always hungry"; "Does food get stuck in your throat?", "No, it never happens". To the question "Do you sometime cough when drinking?" he replied, "No, never". Unlike paresis or dysarthria, which are always present, responding to questionnaires about one's own dysphagia requires the testee to rely on their memory of the symptom. However, Aldo was often confronted by his wife when he was coughing and choking whilst eating or drinking. He denied his dysphagia even when facing it. For example, his wife reported a recent argument they had, as he was gasping for air and almost suffocating after having swigged out of a bottle of water, a habit he was advised to avoid on numerous occasions. Pressed by his wife, disquieted by his irrepressible coughing, he denied any problems maintaining that it was normal to cough sometimes.

The only item in the questionnaire that he acknowledged as a potential problem was having a hoarse voice after eating, which paradoxically is a symptom rarely noticed by dysphagics.

He refused any help to improve his swallowing difficulties with either food or liquids, and never implemented any strategy to avoid possible suffocation. Spontaneously, he often complained of his language deficits, but never of his swallowing problems. Aldo kept denying his swallowing problems even after he was alerted to them by doctors, nurses, speech and physio therapists as well as by his relatives, nor did he acknowledge them after he was shown the outcome of the relevant instrumental diagnostic procedures. His denial for his swallowing difficulties did not change after the training, and he refused to adopt compensatory techniques.

Two years later his conditions further deteriorated yet his anosognosia for his dysphagia remained, whereas he continued to be overtly apprehensive about his dysarthria for which he sought insistent advice.

Dysphagia is not rare in Parkinson-like diseases (Suttrup & Warnecke, 2016; Simons 2017) and can be denied by the patients (Bayés-Rusiñol et al., 2011). Dysphagia has also been described in PSP (Müller et al., 2001). However, in all the cases one can glean from the literature, patients were asked to evaluate their swallowing problems before being confronted with objective assessments or

simultaneously to them. Therefore, their denial has been interpreted as due to inaccurate perception of swallowing abilities (Ding & Logemann, 2008).

The case of Aldo shows that anosognosia for dysphagia can manifest also in the face of clear evidence of the problem and repeated advice from the caring professionals and relatives, to the point of hampering training and resulting in the refusal of helping strategies. Aldo's denial of his very severe dysphagia was not part of a general picture of unawareness of his own deficits, as testified by his preoccupation for his dysarthria and his insistence in being treated for it, even if his articulatory deficits were far less severe and limiting in his daily and social life than his swallowing impairments.

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Table 1. Aldo's neuropsychological assessment four years post-onset.
(References in Supplementary Material).

Test	**Score**
General Cognition	
MMSE	29/30
Milan overall dementia assessment (MODA)	93.9/100
Intelligence and Reasoning	
Verbal Judgements	51/60
Coloured Progressive Matrices (Raven)	27/36
Attention and Executive Functions	
Visual Search	52/60
Trail Making Test B-A	169
Stroop Test errors	9.5*
time	45.5*
Frontal Assessment Battery (FAB)	13/18*
Cognitive Estimation	20/42*
Bizarre responses	7/21*
Card Sorting Test (Wisconsin)	88/128*
Phonological (Letter) Fluency	13*
Semantic (Category) Fluency	28
Language	
Sentences Generation	17/18
Token Test	33/36
Picture Naming	43/48
Memory	
Prose memory Immediate recall	7.9/8
Delayed recall	8/8
Pair Associate Learning	12.5/22.5
Digit Span	5
Spatial (Corsi) Span	5
Complex Figure Recall (Rey)	9/36
Perception	
Complex Figure Copy (Rey)	27/36*
Praxis	
Object use	14/14
Oro-facial praxis	19/20
Gesture Imitation Right arm	72/72
Left arm	71/72
Mood	
Zung Depression Scale	54/80*#

* = Performance below cut-off
\# = Mild depression

SUPPLEMENTARY MATERIALS

References for the neuropsychological tests administered (see Table 1).

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